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Case Report

Successful arterial switch operation for transposition of the great arteries with intact ventricular septum and congenital diaphragmatic hernia: A case report

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ABSTRACT

We report performing a successful arterial switch operation on a full-term male infant with transposition of the great arteries and intact ventricular septum associated with a congenital diaphragmatic hernia. The patient developed severe persistent pulmonary hypertension after successful repair of a left congenital diaphragmatic hernia that restricted pulmonary blood flow into the hypoplastic left lung, and subsequent unilateral pulmonary congestion of the unaffected right lung occurred because of the hemodynamics generated by the transposition of the great arteries. Intravenous epoprostenol with nitric oxide inhalation and mechanical hypoventilation effectively controlled pulmonary vascular resistance before the arterial switch operation was performed.

<**Learning objective:** The outcome of this case suggests that administration of intravenous epoprostenol with nitric oxide inhalation effectively treats persistent pulmonary hypertension of the newborn. Moreover, the hemodynamics associated with transposition of the great arteries requires controlled mechanical hypoventilation to effectively treat pulmonary congestion.>

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Introduction

Perioperative mortality is 2–5% for newborns undergoing arterial switch operations (ASOs) to treat transposition of the great arteries with intact ventricular septum (TGA/IVS) [1]. In contrast, neonatal mortality increases to 50% for neonatal patients with TGA when cases are complicated by persistent pulmonary hypertension of the newborn (PPHN) [2]. Moreover, prognosis is poor when patients present with congenital diaphragmatic hernia (CDH) [3,4], because patients with CDH often suffer from respiratory distress as well as the severest form of PPHN associated with unilateral pulmonary hypoplasia even after successful repair [5]. Here, we report a patient who underwent a successful ASO to treat TGA/IVS with PPHN associated with CDH after effective control of pulmonary vascular resistance.

Case report

A full-term male infant (birth weight: 2790 g) was transferred 3 h after birth to a tertiary medical center because of respiratory distress. Echocardiography revealed TGA/IVS, patent ductal arteriosus, and patent foramen ovale. Moreover, chest radiography revealed an associated left CDH (Fig. 1A). Arterial oxygen saturation was 50% after tracheal intubation and respiratory management using high-frequency oscillating ventilation. Intravenous lipo-prostaglandin (5 ng/kg/min) was initiated to secure ductal patency. The patient remained hypoxic despite successful repair of the CDH 8 h after birth and was therefore transferred to our pediatric cardiac unit. Although balloon atrial septostomy (BAS) was successfully performed on day 1, his oxygen saturation remained at approximately 60%. PPHN was suspected because chest radiography revealed that both lungs were hyperlucent (Fig. 1B), and echocardiography showed a right-to-left shunt at interatrial and ductal levels with no obstructive lesions detectable in the left side of the heart.

Intravenous prostaglandin was discontinued to force the blood into pulmonary arteries by restricting the ductal right-to-left shunt. Nitric oxide (NO) (20 ppm) inhalation was administered to decrease the pulmonary vascular resistance, but the oxygen saturation

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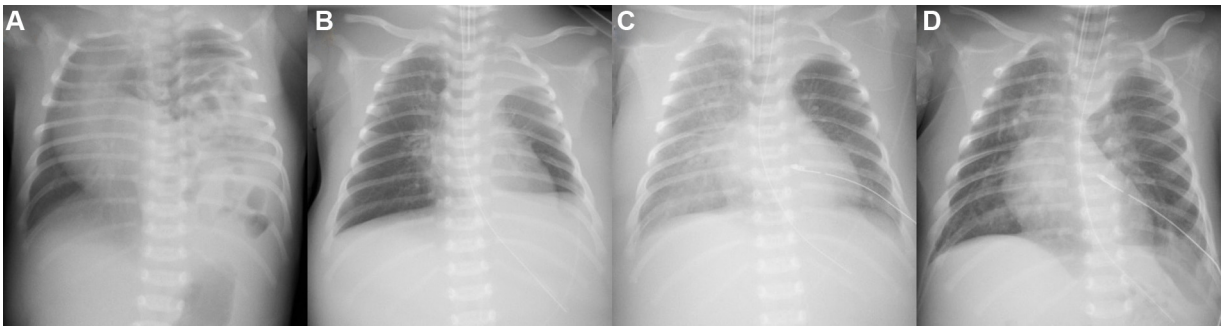

Fig. 1. Chest radiography. (A) Day 0: Left diaphragmatic hernia with all of the small intestine and the right half of the colon prolapsing into the left pleural cavity, displacing the left lung cranially. (B) Day 2: Both lungs were still hyperlucent after the repair of congenital diaphragmatic hernia and balloon atrial septostomy, and the left lung appeared immature. (C) Day 11: An unaffected right lung became congested. A chest tube was placed in the left thoracic cavity. (D) Day 22: Right lung congestion improved following mechanical hypoventilation, and left lung volume increased.

remained at approximately 70%. Then, administration of intravenous epoprostenol (2 ng/kg/min) was initiated on day 2, which immediately increased his oxygen saturation to 80%. Deep sedation and neuromuscular blockade were administered to stabilize oxygenation. Two weeks later, chest radiography and echocardiography revealed increasing pulmonary vascularity and continuous left-to-right ductal flow, respectively. However, the unaffected right lung became congested, although the affected left lung was immature (Fig. 1 C). No obstructive lesions in the pulmonary arteries and veins were detected using echocardiography. To restrict pulmonary blood flow, intravenous epoprostenol and inhaled NO were discontinued on days 13 and 14, respectively; however, the previously unaffected right lung remained congested. Further, bilious gastric aspirates and distended abdomen suggested potential risk of bowel complications after an immediate ASO. Therefore, controlled mechanical ventilation was performed to maintain high arterial CO_2 partial pressure (up to 55 cm H_2O), which increased the pulmonary vascular resistance and regulated the pulmonary blood flow into the congested right lung (Fig. 1D). ASO was successfully performed on day 23 without complications following the improvement of bowel status and the growth of the hypoplastic left lung. Mechanical ventilation was discontinued on day 30. The patient was discharged with no medication on day 54.

Discussion

We report here a successful ASO for TGA/IVS with PPHN associated with left CDH. The clinical course provides important insights into the strategy for treating patients with TGA with severe pulmonary complications.

Although patients with CDH suffer the severest form of PPHN associated with unilateral pulmonary hypoplasia [5], postnatal growth and remodeling of the lungs occur after CDH repair [6]. Considering that performing an ASO on patients with TGA with PPHN during the unstable early neonatal period may increase the risk of mortality [1], we decided to wait for the growth of the affected left lung and for improved nutrition of the patient. However, we encountered two major problems during the treatment.

The primary problem was the association of PPHN. Hypoxia persisted after successful repair of the CDH on day 0, although BAS was performed to improve interatrial blood mixing on day 1. The diagnosis of PPHN was supported by the presence of interatrial and ductal right-to-left dominant shunts, which caused reverse differential cyanosis. Therefore, we discontinued the administration of intravenous prostaglandin to prevent the collapse of the pulmonary circulation [7]. Oxygenation was further stabilized by deep sedation, neuromuscular blockade, inhaled NO, and intravenous epoprostenol. Administration of intravenous epoprostenol was particularly effective. To the best of our knowledge, there are no published reports on the successful use of intravenous epoprostenol to treat patients with TGA, although inhaled NO, extracorporeal membrane oxygenation, and bosentan are beneficial [2,8].

Asymmetrical pulmonary vascular resistance caused the second problem. The unaffected right lung became congested with high pulmonary blood flow concurrent with the improvement of PPHN, although the affected left lung remained immature with low vascularity. Beals et al. [6] reported postnatal remodeling of the lungs in patients with CDH, and showed that the unaffected right lung undergoes a greater amount of remodeling than the affected left lung. Because an echocardiography showed no obstructive lesions in the pulmonary arteries and veins, we assumed that pulmonary vascular resistance was unbalanced. Growth of the affected lung, stabilization of pulmonary vascular resistance, and improved nutrition required more time. Therefore, we considered administering mechanical hypoventilation as a reasonable measure to restrict the right pulmonary blood flow. For example, Barnea et al. [9] first showed that controlled mechanical hypoventilation successfully manipulates pulmonary vascular resistance in a univentricular model, suggesting its application in TGA/IVS when systemic and pulmonary circulations are parallel. By raising pulmonary vascular resistance, this procedure may help in maintaining adequate pulmonary arterial pressure to ensure sufficient left ventricular mass to sustain systemic pressure following ASO [10]. In patients with TGA, when pulmonary vascular resistance of the unaffected lung decreases, imbalanced resistance develops within the lung, and the unaffected right lung will suffer from high pulmonary blood flow. Aggressive treatment of PPHN and the management of subsequent unilateral pulmonary congestion by increasing the pulmonary vascular resistance through the ventilator setting was likely the key to the success of the ASO.

A limitation of this case report was that the poor condition of the patient prevented us from confirming the basis of these responses using cardiac catheterization. However, we are confident that the diagnosis of PPHN was supported by the findings of chest radiography and echocardiography studies as well as by the subsequent decrease in cyanosis after treatment with epoprostenol and inhaled NO. The imbalance of pulmonary vascular resistance was also strongly suggested by the laterality of pulmonary vasculature revealed by the chest radiography as we did not detect obstructive lesions in the heart by echocardiography as mentioned earlier, which was further supported by the clinical improvement of pulmonary congestion following hypoventilation therapy.

In conclusion, treatment with intravenous epoprostenol and NO inhalation was effective in PPHN, and mechanical hypoventilation therapy managed subsequent unilateral pulmonary congestion even under the unfavorable hemodynamic conditions of TGA.

Conflicts of interest

The authors declare no conflicts of interest.

Acknowledgments

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